Anesthetic and Airway Management in a Pediatric Patient with Morquio Syndrome: A Case Report

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ABSTRACT

Morquio syndrome is a subtype of mucopolysaccharidoses, wherein the accumulation of glycosaminoglycans (GAGs) in various organ systems lead to alteration of anatomy and physiology. Most prominent features are extensive bony abnormalities, which normally require surgical correction. This paper reports the case of a 7-year-old child with Morquio syndrome who successfully underwent correction of genu valgum under general endotracheal anesthesia via asleep induction and videolaryngoscopy, with supplemental peripheral nerve block. The precautions and anesthetic care done to ensure a safe procedure are discussed, especially with anticipation of a possible difficult airway.

Keywords: Morquio syndrome, mucopolysaccharidosis, difficult airway management, anesthesia

INTRODUCTION

Morquio syndrome, also known as Mucopolysaccharidosis type IV (MPS IV), is a rare autosomal recessive birth defect that is estimated to occur in one of every 40,000 to 200,000 births.[1] It results from mutations in the gene encoding galactosamine-6-sulfatase (Type A) or beta-galactosidase (Type B), consequently leading to accumulation of keratan sulfate and chondroitin-6-sulfate in multiple organs.[2] The buildup of mucopolysaccharides in various organ systems leads to short stature, spinal deformity, odontoid hypoplasia, macroglossia, corneal opacities, pectus carinatum, kyphoscoliosis, cardiac abnormalities, hepatomegaly, hearing loss, and dental abnormalities.[3] Of important note is that patients with MPS IV have normal intelligence.[4] The greatest challenge to the anesthesiologist is possible difficult airway from a short neck and possible infiltration of pharyngeal and laryngeal structures, and even the trachea, with GAGs, causing narrowing of the airway which cannot be ascertained during physical examination. Clinical presentation of patients with MPS IV may have severe or attenuated forms, depending upon the remaining residual enzyme activity, and a thorough understanding of all these anatomic and physiologic changes and their extent is paramount in overcoming the anesthetic challenges that they present with.

CASE DESCRIPTION

A 7-year-old male was scheduled for elective bilateral medial hemiepiphysiodesis after complaints of knee pain from progressive genu valgum. He had a complete cessation of growth at 4 years of age, and was diagnosed with Morquio

Poster presentation – 23rd ASEAN Congress of Anesthesiologists 2023 (ACA 2023) in conjunction with the 98th Annual Scientific Meeting of the Royal College of Anesthesiologists of Thailand, September 1-3, 2023, Royal Orchid Sheraton Hotel and Towers, Bangkok, Thailand.

eISSN 2094-9278 (Online)
Published: May 30, 2024
https://doi.org/10.47895/amp.v58i9.8284

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syndrome Type A after undergoing gene testing. The patient weighed 12.8 kg and was 93 cm tall (BMI 15.1 kg/m^2). Results of liver function test, renal function test, echocardiogram, and echocardiography previously done were all within normal. Notable on history was snoring and predilection for a side lying position during sleep, indicative of partial airway obstruction. On physical examination, he was active, playful, and was developmentally at par in all domains. Preoperative vital signs include a blood pressure of 90/60 mmHg, heart rate of 108 beats per minute, respiratory rate of 22 breaths per minute, temperature of 36.7˚C and an oxygen saturation of 100% at room air. Prominent physical features include a short stature with dolichocephaly, short neck, pectus carinatum, and genu valgum. Airway assessment showed an adequate mouth opening and thyromental distance, a Mallampati score of 2, and no limitation in neck extension. There were no cardiac murmurs appreciated on auscultation, and breath sounds were clear. The patient's parents were apprised preoperatively of the anesthetic plans and risks, including possible difficulty in securing the patient's airway.

Preparations for anesthesia included a complete airway set-up consisting of varying sizes of endotracheal tubes (ETT), supraglottic airways (SGA), oral and nasal airways, bougies, intubating stylets, and suction catheters. Apart from the conventional laryngoscope, a videolaryngoscope with a pediatric D-blade and MAC 2 blade was readied. A pediatric fiberoptic bronchoscope was also placed on standby.

Once at the operating room, standard noninvasive monitors were attached to the patient, including a noninvasive blood pressure monitor, 5-lead ECG, pulse oximeter, temperature probe, and a precordial stethoscope. Rolled linen was placed under the patient's shoulders to avoid excessive neck flexion while in the supine position as seen in Figure 1. The patient was then premedicated with Midazolam 0.05 mg/kg IV and Atropine 0.01 mg/kg IV. Preoxygenation was done, and the following medications were administered for the induction of anesthesia: Ketamine 1 mg/kg IV, Remifentanil target-controlled infusion (TCI) at 3 ng/ml and Propofol 2 mg/kg IV slowly titrated to achieve hypnosis. The goal was to maintain the patient's spontaneous respirations until the ability to ventilate was ensured. Once a satisfactory mask ventilation was established, the patient was paralyzed using Rocuronium 0.6 mg/kg IV. Sugammadex 8 mg/kg IV was already aspirated and available for use in case a cannot ventilate, cannot intubate scenario ensues.

The initial attempt to secure the airway was done using videolaryngoscopy with a pediatric D-blade to keep the neck in a neutral position. There was ease in inserting the blade and visualizing the glottic opening. The videolaryngoscopy view is shown in Figure 2. A size 5.0 cuffed ETT was inserted smoothly after one attempt, secured at level 15, and bilateral clear and equal breath sounds were noted thereafter. An adequate depth of anesthesia was maintained using Sevoflurane and Remifentanil TCI. Given the restrictive lung physiology resulting from the patient's thoracic bony

Figure 1. Photo of the patient post-induction of anesthesia showing pectus excavatum and short neck.

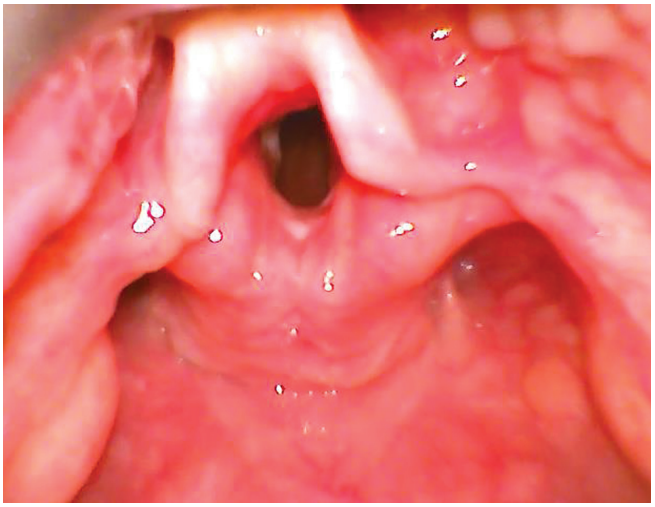

Figure 2. CMAC videolaryngoscopy view using pediatric D-blade showing a clear visualization of the glottic opening.

deformities, mechanical ventilation was set at pressure control ventilation with positive end-expiratory pressure (PEEP) of 3 cm H_2O, inspiratory pressure of 12-13 cm H_2O, and respiratory rate of 26-27 breaths per minute. End-tidal carbon dioxide ($ETCO_2$) was maintained at 35-40 mmHg throughout the procedure.

After securing the airway, a second intravenous access was secured. Ultrasound-guided bilateral femoral nerve and popliteal sciatic nerve blocks were then done to augment analgesia. A sensory dose of Bupivacaine 0.2% + epinephrine 1:400,000 4.5 ml per side was administered for both blocks, visualizing the spread of the local anesthetic around the target nerves. Notable was a more proximal branching of the sciatic nerve into the common peroneal and tibial nerve on the left popliteal area compared to the right side. Total dose of

Bupivacaine given was 36 mg, well within the toxic dose of 38.4 mg for the patient.

Total surgery time was 1 hour and 42 minutes. A tourniquet was used to decrease blood loss from the procedure. A forced-air warming blanket was applied to maintain the patient normothermic intraoperatively at 36-37°C. The patient was extubated fully awake at the end of the surgery after reversal of neuromuscular blockade, without any complications. He was kept in a recovery position while at the Post-Anesthesia Care Unit (PACU) to keep the airway clear and open, and monitored for desaturations.

After ensuring good oral intake and adequate pain control, the patient was discharged well on the first post-operative day.

DISCUSSION

Many patients with Morquio syndrome will require surgery throughout their lifetime to improve their function and comfort, and an even greater number will need anesthesia if those for sedation during imaging studies is included.[5] A careful and thorough pre-operative evaluation of the patient and an exhaustive planning for the procedure is key to ensuring good outcomes.

Common features of patients with MPS IV include a short stature from a shortened neck and trunk, protuberant chest (pectus carinatum), deformed and misshapen long bones, extreme short stature, and knock-knee deformity (genu valgum). They may also have kyphoscoliosis and joint laxity. Odontoid hypoplasia is also common, leading to atlanto-axial instability and C1-C2 subluxation. In the respiratory system, they are at risk for restrictive pulmonary disease, reduced alveolar capacity, and recurrent infections due to thoracic deformities. They may also have a limited mouth opening and progressive narrowing of upper airway related spaces, causing sleep apnea or disordered breathing. In terms of the cardiovascular system, GAGs may accumulate in the coronary arteries, heart valves, and myocardium, causing dysfunction. MPS IV patients are at also at risk of multilevel spinal canal stenosis and cord compression, leading to neurologic deficits.[6]

From the knowledge of the affectation of Morquio syndrome and the physical examination of the patient, a possible difficult airway was the primary concern for this case. The patient had a short neck which may limit neck movement and make visualization of the glottic opening challenging during laryngoscopy.[7] Beyond what can be assessed during routine physical examination, GAGs may also infiltrate pharyngeal and laryngeal structures, causing narrowing of the upper airway, and this may be suggested by the patient's observed sleep-disordered breathing. Additionally, keratan sulfate, the predominant GAG in MPS IV, has a predisposition to accumulate in the trachea, especially the hyaline cartilage of the anterior tracheal rings, leading to tracheal narrowing and tracheomalacia.[6] Anesthesiologists must be prepared to manage all these potential airway challenges if anesthesia is desired for any procedure.

The foremost challenge in securing the airway of this patient is establishing effective mask ventilation, which will determine the medications to be given and mode of induction of anesthesia and intubation. If mask ventilation can be established, a neuromuscular blocker may be given to facilitate intubation; otherwise, spontaneous ventilation must be preserved. Ventilation via a supraglottic airway may also be attempted.

After effective mask ventilation was established, it was crucial to use an easily reversible neuromuscular blocker—for this case Rocuronium with Sugammadex on standby for immediate reversal of paralysis—in case sudden difficulty in mask ventilation ensues. The initial method of choice for securing the airway was through videolaryngoscopy to avoid excessive neck movement[8], with the use of a pediatric D-blade providing an added benefit of improved glottic visualization and quick intubating times[9,10]. The former is important since patients with MPS IV have a hypoplastic odontoid process which leads to atlanto-axial instability and places the patient at an inherent risk of quadriparesis. If the patient's airway cannot be intubated through traditional means, back-up plan was via a fiberoptic bronchoscope, or if still impossible, the alternate plan was to secure the airway via a supraglottic airway.

Various airway equipment and modalities were thus organized for use for this case. Computed ETT size based on age was 5.0 cuffed, but sizes 3.5-5.0 uncuffed and 3.0-5.5 cuffed tubes were placed on standby in case narrowing in any part of the airway is encountered. Intubating stylets and bougies corresponding to the different tube sizes were prepared as well. Given the critical role of a supraglottic airway both as means for ventilation and as a definitive airway, both LMA supreme and i-gel sizes 1.5 and 2 were prepared for the case.

A sensory block of the bilateral femoral and popliteal sciatic nerves was done to provide intraoperative and post-operative pain relief, as part of a multimodal analgesia technique. It was essential to decrease the post-operative opioid requirement in this patient with an already obstructed airway during sleep, as opioids can worsen the airway obstruction and decrease the central respiratory drive.[11] The need for an ultrasound-guided approach to peripheral nerve block is highlighted during the case, with noted anatomic variations in the level of the bifurcation of the sciatic nerve in the right and left lower extremities. Perhaps a landmark-based approach to peripheral nerve blocks may not always precisely locate the target nerves in patients with Morquio syndrome, although this may need further studies.

Other options for the anesthetic management of a patient with MPS IV undergoing lower extremity surgery would have been regional anesthesia. In other patient profiles, a surgical block of the bilateral femoral and popliteal sciatic nerves may have possibly sufficed for the surgery,

and could have avoided the tedious preparations for airway management. However, the psychological development of this child precludes this option. A pure surgical block would have necessitated moderate to deep sedation for this patient, risking loss of the airway anytime during the sedation in an uncontrolled setting, which may consequently lower the success rate in securing the airway. The dose of local anesthetic required for a surgical peripheral nerve block would also be higher, and could go beyond the safe dose prescribed for the patient's weight. Neuraxial anesthesia was dismissed as an option for this patient due to reports of spinal cord injury in patients with MPS IV following neuraxial anesthesia.[12] Although there were success stories[13], ultrasound-guided peripheral nerve block was deemed as a safer option.

CONCLUSION

Patients with Morquio syndrome have a multisystemic affectation with varying presentation and severity, which imposes challenges to the anesthesiologist. A thorough pre-anesthetic evaluation is of utmost importance to come up with an individualized anesthetic plan. Careful planning and preparation, which includes consideration for the psychological development of pediatric patients and communication of risks and benefits to the child's guardians, are optimal for successful management. Anesthetic risks should be communicated to the healthcare team as well. With an anticipated difficult airway, a spontaneous respiration is usually maintained for these patients until the ability to mask ventilate or a definite airway is established. Visualization of the glottic opening via videolaryngoscopy using a pediatric D-blade remains a viable option for securing the airway in patients with MPS IV while maintaining neutral neck positioning.

Informed Consent

Informed consent was obtained from the patient's guardian for the publication of the patient's clinical information and images.

Statement of Authorship

Both authors certified fulfillment of ICMJE authorship criteria.

Author Disclosure

Both authors declared no conflicts of interest in preparing this article.

Funding Source

No specific grant from any funding agency in the public, commercial or not-for-profit sectors was received in writing this paper.

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